

## CLAIMS

1. A human gene for synthesizing an enzyme catalyzing human *N*-linked sugar chain synthesis, which is homologous with a gene of an enzyme catalyzing *N*-linked sugar chain synthesis in yeast endoplasmic reticulum, and is capable of complementing the function of said gene for a deletion yeast strain of said gene.

2. The human gene according to claim 1, wherein the enzyme catalyzing human *N*-linked sugar chain synthesis is a glycosyltransferase.

3. A gene which encodes the amino acid sequence represented by SEQ ID NO:2, 4, 6, 8 or 10 or a protein which comprises an amino acid sequence in which one or more amino acids in the amino acid sequence represented by SEQ ID NO:2, 4, 6, 8 or 10 are deleted, substituted or added.

4. A gene which comprises the nucleotide sequence represented by SEQ ID NO:1, 3, 5, 7 or 9.

5. An agent for diagnosing or treating human congenital disorders of glycosylation syndrome, which comprises using the gene encoding the amino acid sequence according to claim 3 or the gene represented by SEQ ID NO:1, 3, 5, 7 or 9.

6. A recombinant vector which is integrated with the gene according to any one of claims 1 to 3.

7. A transformant which is transformed by the recombinant vector according to claim 6.

8. A process for producing an enzyme catalyzing human *N*-linked sugar chain synthesis, which comprises culturing the transformant according to claim 7 in a culture, and collecting the enzyme catalyzing human *N*-linked sugar chain synthesis from the culture.

9. A method for synthesizing a human *N*-linked sugar chain, which comprises using the enzyme according to claim 8.